Lack of Torsion in the Type-III Duane Retraction Syndrome (DRS-III) Affected Left Eye During Dix -Hallpike Test

Ajay Kumar Vats[1]

[1]Department of Neurology, Chaudhary Hospital and Medical Research Centre Private Limited, Hiran Magri, Udaipur, Rajasthan, India

Address for correspondence Ajay Kumar Vats, MBBS, MD, DM, Department of Neurology, Chaudhary Hospital and Medical Research Centre Private Limited, 472-473, Sector 4, Hiran Magri, Udaipur, Rajasthan 313002, India (e-mail: vatsneuro@gmail.com).

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Abstract

Background Duane retraction syndrome (DRS) is a congenital cranial dysinnervation disorder (CCDD) of ocular movements, characterized by deficits in horizontal duction associated with narrowing of palpebral fissure, retraction of eye globe on attempted adduction and occasionally accompanied by upshoot or downshoot of the eye globe. It is caused by congenital absence of sixth cranial nerve, which results in fibrotic changes in the extraocular muscles leading to an abnormal ocular motility--a concept known as CCDD. Depending on whether only abduction/adduction or both are affected, DRS has been classified into three types designated as type-I, type-II and type-III. The torsional movements of the affected eye in DRS have not been reported to be deficient hitherto, which could be due to difficulties in the routine bedside evaluation of such movements.

Case Presentation An unusual case of a patient of left unilateral type-III DRS is reported, who presented with a short history of vertigo on getting up from supine to sitting position and on assuming right lateral recumbent position. The diagnostic right Dix-Hallpike test (DHT) revealed upbeating torsional geotropic positioning nystagmus in the normal right eye and upbeating positioning nystagmus without torsional component in the abnormal left eye and this clinical finding was video recorded. **Conclusion** The observed lack of incyclotorsion of the left eye, affected with DRS-III during right Dix-Hallpike positioning, is primarily due to the absence of initial slow-phase excyclotorsional component. If the slow phase of VOR does not occur, then the fast-phase VOR, which is a refixation saccade, will be lacking too. An anastomosis, either in the lateral wall of the cavernous sinus or within the orbit, between the trochlear nerve and fibers of the oculomotor nerve can lead to simultaneous co-contraction of the inferior and superior oblique muscles. This is the most probable explanation for such finding of asymmetrical absence of torsional component in the left eye affected by DRS-III, during right Dix -Hallpike positioning. Thus, the recording of eye movements (voluntary and involuntary) opened a window into the brain to conceptualize neural and mechanical factors influencing the human eye movements.

Keywords
- cranial dysinnervation disorder
- Dix–Hallpike test
- incylotorsional
- excyclotorsional
- positioning nystagmus
- geotropic

Introduction

Duane's retraction syndrome (DRS) is a congenital cranial dysinnervation disorder (CCDD) of ocular movements, characterized by deficits in horizontal duction, which is associated with narrowing of palpebral fissure, retraction of eye globe on attempted adduction, and occasionally accompanied by upshoot or downshoot of the eye globe. DRS is now widely accepted to be caused by congenital absence of sixth

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cranial nerve, which results in fibrotic changes in the extraocular muscles leading to abnormal ocular motility, a concept known as CCDD.[1] The developmental anomaly of one or more cranial nerves may either result in congenital dysinnervation of the cranial muscles or else latter may secondarily develop aberrant innervations from other cranial nerves. The electromyographic (EMG) studies in patients with DRS have demonstrated cocontraction of medial and lateral recti muscles and the adduction of affected eye causes its retraction with narrowing of palpebral fissure.[2] The upshoots and downshoots are abrupt vertical deviations occurring in the affected eye of the patients suffering from DRS during adduction and are putatively attributed to mechanical and innervational causes. The sideslip of the tight lateral rectus muscle when the eye globe moves above or below the horizontal plane in adduction is believed to be the mechanical reason for upshoots and downshoots and it has been termed as "bridle effect."

Based on the degree of asymmetry between the limitation of adduction and abduction, DRS has been classified into three types as follows.[3]

Type-I: characterized by pronounced limitation or absence of abduction, normal or only slightly impaired adduction, narrowing of the palpebral fissure and retraction on adduction, and widening of the palpebral fissure on attempted abduction.

Type-II: characterized by limitation or absence of adduction with exotropia of the affected eye, normal or slightly impaired abduction, narrowing of the palpebral fissure, and retraction of the globe on attempted abduction.

Type-III: characterized by a combination of limitation of both abduction and adduction, retraction of the globe, and narrowing of the palpebral fissure on attempted adduction.

I am reporting here a very unusual case of a patient with left unilateral type-III DRS, who presented with 5-day history of vertigo on getting up from supine to sitting position and on assuming right lateral recumbent position. The diagnostic right Dix–Hallpike test (DHT) revealed upbeating counterclockwise torsional geotropic positioning nystagmus in the normal right eye and upbeating positioning nystagmus without torsional component in the abnormal left eye and this observation was video recorded.

Case Description

A 21-year-old female patient presented with 5-day history of vertigo on getting up from supine to sitting position and on assuming right lateral recumbent position. There was no history of staggering, diplopia, dysarthria, difficulty in swallowing, hiccups, drooping of upper eyelids, facial, or limb weakness. She informed that her left eye had a problem in movement and alignment since childhood. The neurological examination of cranial nerves revealed abnormal extraocular movements in the form of limitation of abduction, as well as adduction, of the left eye and attempt to adduct left eye resulted in retraction, as well as downshoot, of the left eye, which was consistent with diagnosis of type-III unilateral DRS (►**Video 1**). The power was grade 5/5 in all four limbs with normal deep tendon reflexes and bilateral plantar reflexes were flexor. The examination of cerebellar system revealed no spontaneous or gaze evoked nystagmus and no appendicular or axial incoordination was observed. The otoneurological examination revealed normal vertical and horizontal saccadic and smooth pursuit eye movements of the right eye. In the left extreme gaze, the normal right eye was fully adducted and the affected left eye revealed marked limitation of abduction (►**Fig. 1**). In the extreme right gaze during smooth pursuits, the normal right eye was fully abducted and the affected left eye revealed marked limitation of adduction with retraction and downshoot (►**Fig. 2**). The head impulse test was normal in the right eye and uninterpretable in the left eye. The DHT on the right side was performed by making the patient sit on the examination couch with both lower limbs placed along the long axis of the couch. A 4-inch thick pillow was placed behind her buttocks to be used as vantage point instead of using the end edge of the couch, during the test. Her head was rotated 45 degrees to her right in the yaw plane and she was taken to supine position so that her head got extended 20 degrees as she was laid. Right DHT elicited a positioning nystagmus, which was upbeating and excyclotorsional in the normal right eye and upbeating in diseased left eye (►**Video 2**).

Video 1

The video shows examination of smooth pursuits, which reveal deficiency of adduction, as well as abduction, of the left eye. The abduction of left eye is more severely restricted compared with adduction. There is narrowing of left palpebral fissure and downshoot of the left eye on adduction. The vertical smooth pursuit movements are normal. The left eye shows deficiency of adduction on convergence. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0040-170807.

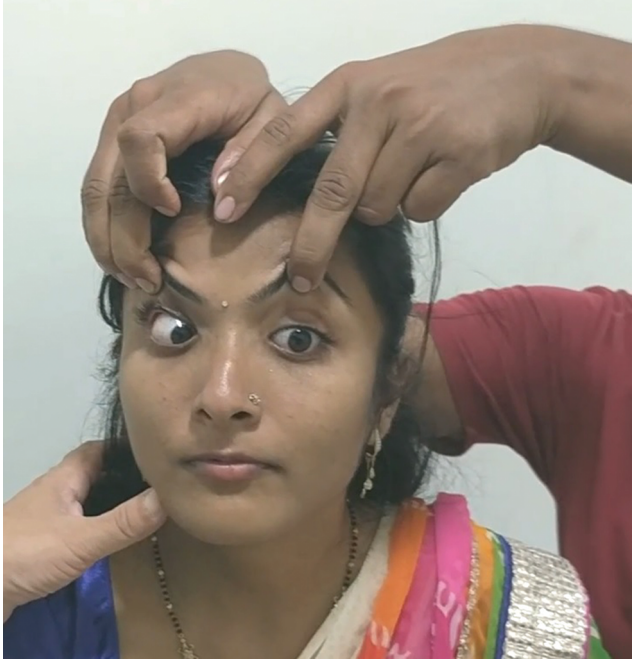

Fig. 1 In the left extreme gaze while testing smooth pursuits, the normal right eye was fully adducted and the affected left eye reveled marked limitation of abduction.

Fig. 2 In the extreme right gaze during smooth pursuits, the normal right eye was fully abducted and the affected left eye revealed marked limitation of adduction with retraction and downshoot.

Video 2

The Dix–Hallpike test on the right side was performed by making the patient sit on the examination couch with both lower limbs placed along the long axis of the couch. A 4-inch thick pillow was placed behind her buttocks to be used as vantage point instead of using the end edge of the couch, during the test. Her head was rotated 45 degrees to her right in the yaw plane and she was taken to supine position so that her head got extended 20 degrees as she was laid. Right Dix–Hallpike test elicited a positioning nystagmus, which was upbeating and excyclotorsional in the normal right eye and upbeating without torsional component in the diseased left eye. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0040-170807.

Video 3

The Dix–Hallpike test on the right side was performed by making the patient sit on the examination couch with both lower limbs placed along the long axis of the couch. A 4-inch thick pillow was placed behind her buttocks to be used as vantage point instead of using the end edge of the couch, during the test. Her head was rotated 45 degrees to her right in the yaw plane and she was taken to supine position so that her head got extended 20 degrees as she was laid. Right Dix-Hallpike test elicited a positioning nystagmus, which was upbeating and excyclotorsional in the normal right eye and upbeating without torsional component in the diseased left eye. The initial slow-phaseVOR, which in downbeating in both eyes, incyclotorsional in right eye and lacking excyclotorsional component in DRS-III affected left eye is better appreciated with video speed slowed to 25% of the original. DRS, Duane's retraction syndrome; VOR, vestibuloocular reflex. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0040-170807.

Note: The speed of video was decreased to 25% of the original by using the online web url: https://clideo.com/change-video-speed.

The patient was treated with Epley's maneuver and she had complete relief in vertigo.

Discussion

The slow phase of the vestibuloocular reflex (VOR) stabilizes the gaze in space by moving the eyes in direction opposite to the head movement, while the fast phase redirects gaze at high speeds in the direction of head velocity. In a patient with right posterior semicircular canalolithiasis without CCDD, the right Dix–Hallpike positioning aligns the right posterior semicircular canal vertically, causing ampullofugal movement of endolymph, and otoconial debris under the effect of gravitational force. The stimulation of right semicircular canal strongly activates a pair of ocular muscles, one in each eye, aligned with its plane of action and concurrently inhibits the antagonist pair. During the right Dix–Hallpike positioning in a patient with right posterior canalolithiasis, the right superior oblique (SO) and left inferior rectus (IR) receive excitatory impulses from the right semicircular canal, while the antagonistic pair of muscles, the right inferior oblique (IO), and left superior rectus (SR) simultaneously receive the inhibitory impulses. The right Dix–Hallpike positioning results in slow-phase VOR which is downbeating in both eyes with incyclotorsional component in the right eye and excyclotorsional in the left eye (►**Fig. 3**). Therefore, the fast-phase VOR which is a refixation saccade (clinically appreciated as positional nystagmus), will be upbeating in both eyes with excyclotorsional component in the right eye and incyclotorsional component in the left eye. In this patient with DRS-III, during the right Dix–Hallpike positioning, the slow phase of the VOR appears as downbeating plus incyclotorsional in the normal right eye followed by the corrective fast-phase VOR (►**Video 3**). Thus, the positional nystagmus

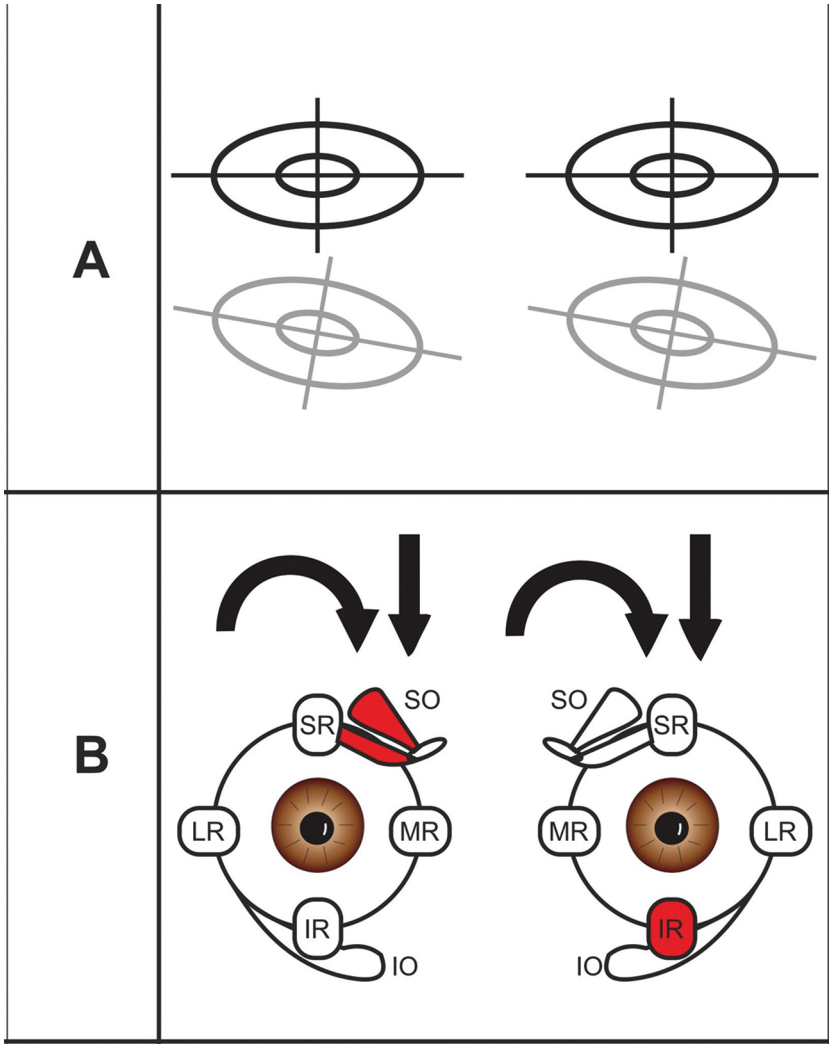

Fig. 3 (**A**) Position of eyes at rest (black colored) and during slow-phase VOR (gray colored) of the right Dix-Hallpike positioning in patient with right posterior canalolithiasis without DRS-III. (**B**) The right posterior semicircular canal excitatory projections to right superior oblique and left inferior rectus muscles (colored red) produce slow phase VOR during right Dix-Hallpike positioning, which is downbeating for both eyes; incyclotorsional for right eye and excyclotorsional for left eye. VOR, vestibuloocular reflex; DRS, Duane's retraction syndrome; IO, inferior oblique; IR, inferior rectus; MR, medial rectus; SO, superior oblique; SR, superior rectus.

Table 1 Summary of findings of right Dix–Hallpike test in a patient of right PSC-BPPV with left eye affected by DRS-III

Vestibuloocular reflex	Right eye		Left eye	
	Vertical component	Torsional component	Vertical component	Torsional component
Slow phase	Downbeating	Incyclotorsional	Downbeating	Absent
Fast phase (positional nystagmus)	Upbeating	Excyclotorsional	Upbeating	Absent

Abbreviations: DRS, Duane's retraction syndrome; PSC-BPPV, posterior semicircular canal benign paroxysmal positional vertigo.

(fast-phase VOR) in the right eye is upbeating and excyclotorsional. By contrast, in the left eye affected by DRS-III, the slow phase of the VOR appears as downbeating without excyclotorsional component and this is followed by the corrective fast phase, which is upbeating without incylotorsional component (►**Videos 2** and **3**). The result of right DHT in this patient with left eye affected by DRS-III is summarized in the ►**Table 1**. The observed deficiency of VOR slow-phase excyclotorsion followed by lack of corrective fast-phase incylotorsion of left eye affected by unilateral DRS-III, during right Dix–Hallpike maneuver is phenomenal. The absence of slow-phase excyclotorsion and fast-phase incyclotorsion of the VOR in the DRS-III affected left eye during right DHT can be explained with cranial dysinnervation or aberrant innervation. Ramification of branches arising from the inferior division of oculomotor nerves have been demonstrated to penetrate the inferomedial portion of the lateral rectus muscle in the autopsy studies.[4] Aside from these ramifying branches derived from the oculomotor nerve, the rest of the lateral muscle mass

in autopsy reports was found to be poorly innervated and fibrotic, and the medial rectus was normal in size, as well as structure. These autopsy studies in patients with DRS-III have reported hypoplastic abducens nucleus lacking motor neuron cell bodies. Instead several small cell bodies appearing like internuclear neurons have been found but no identifiable intra-axial abducent nerve fibers within the brainstem. At the level of the ciliary ganglion, the oculomotor nuclei and nerves were found to be normal.

By similar analogy, an anastomosis between branches emanating from the inferior division of the oculomotor nerve and the SO muscle within the orbit, can also be expected to cause cocontraction of IO and SO muscles. The lateral wall of the cavernous sinus is another place where trochlear and oculomotor nerves are in proximity. An anastomosis within the cavernous sinus, between trochlear nerve and oculomotor nerve fibers destined to become its inferior division in the orbit, is also expected to cause cocontraction of IO and SO muscles. The alternative explanation for lack of torsion of DRS-III affected left eye could be intraorbital mechanical hindrance. The patient was treated with Epley'smaneuver and she had complete relief in vertigo.

Conclusion

The recording of eye movements (voluntary and involuntary) opens a window into the brain to conceptualize neural and mechanical factors influencing the human eye movements. In the case discussed above, the video recording of induced positioning nystagmus during right DHT, for the evaluation of positional vertigo revealed lack of slow-phase excyclotorsion, as well as fast-phase incyclotorsion in the DRS-III affected left eye, a finding not reported hitherto. The incyclotorsional movement of any eye is executed by the contraction of SO muscle that receives nerve supply from the trochlear cranial nerve arising from the contralateral cerebral hemisphere. The reports of autopsy done in patients of DRS have never reported abnormalities of trochlear nerve (nucleus, intrabrainstem, or extra-axial portion). Therefore, observed lack incyclotorsion of left eye, affected with the DRS-III during Dix–Hallpike positioning, is primarily due to absence of initial slow-phase excyclotorsional component. If slowphase of VOR does not occur, then the fast-phase VOR, which is a refixation saccade, will be lacking too. An anastomosis, either in the lateral wall of cavernous sinus or within the orbit, between trochlear nerve and fibers of oculomotor nerve can lead to simultaneous cocontraction of the inferior and SO muscles. This is the most probable explanation for such finding of asymmetrical absence of torsional component in the left eye affected by DRS-III, during right Dix–Hallpike positioning.

Note

The views expressed in the submitted article are author's own and not an official position of the institution to which author is affiliated.

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Conflict of Interest

None declared.

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References

1 Parsa CF, Grant PE, Dillon WP Jr., du Lac S, Hoyt WF. Absence of the abducens nerve in Duane syndrome verified by magnetic resonance imaging. Am J Ophthalmol 1998;125(3):399–401
2 Breinin GM. Electromyography; a tool in ocular and neurologic diagnosis. II. Muscle palsies. AMA Arch Opthalmol 1957;57(2):165–175
3 Von Noorden GK, Campos EC, eds., Special forms of strabismus: retraction syndrome (Duane syndrome). In: Binocular Vision and Ocular Motility. 6th ed. St. Louis, MO: Mosby Inc.; 2002 458–466
4 Hotchkiss MG, Miller NR, Clark AW, Green WR. Bilateral Duane's retraction syndrome. a clinical-pathologic case report. Arch Ophthalmol 1980;98(5):870–874